Middle aortic syndrome: Surgical treatment in a child with neurofibromatosis

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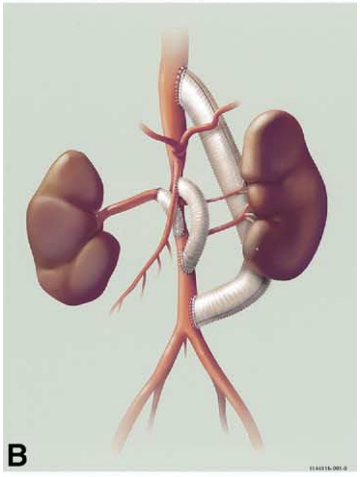
A 9-year-old boy was referred to our institution for management of severe renovascular hypertension (controlled with 6 antihypertensive medications), postprandial abdominal pain, and bilateral lower-limb claudication. He was diagnosed with neurofibromatosis at age 6, manifested by multiple café-au-lait macules (>5cm), axillary and inguinal freckling, and brain lesions on magnetic resonance imaging consistent with neurofibromas. Physical findings included a midepigastric bruit and attenuated lower limb arterial pulses (ankle-brachial index (ABI), $0.67_{[Rt]}/0.7_{[Lt]}$). Digital subtraction angiography revealed segmental narrowing of the abdominal aorta from below the celiac to the inferior mesenteric artery (mean pressure gradient 30 mm Hg) and orificial stenoses (>75%) of the right renal (A) and superior mesenteric arteries (A, *insert*). Investigation excluded a pheochromocytoma. Surgery, through a midline incision, entailed an aortoaortic bypass (14-mm gelatin-coated polyester) from the supraceliac to distal infrarenal aorta, tunneled posterior to the left kidney, and a retrograde bypass to the superior mesenteric (end-to-side) and right renal (end-to-end) arteries, using 6-mm collagen-coated polyester grafts from the infrarenal aorta (B [Cover]). The patient was discharged on the fifth post-operative day on a single antihypertensive medication, with normal pulses and pressure indexes (ABI, $1.07_{[Rt]}/1.09_{[Lt]}$), and a serum creatinine of 0.86. Four months postoperatively the child was normotensive on no drug treatment, free of abdominal pain and claudication, with all of his grafts patent and stenosis-free, as depicted in 3-dimensional contrast computed tomography imaging of the reconstructed aortovisceral circulation (C).



Segmental narrowing of the abdominal aorta, designated middle aortic syndrome (MAS),[1] represents an uncommon variety (0.5%-2%) of aortic coarctations attributable to acquired or congenital etiologies.[2-4] Acquired etiologies include non-specific (Takayasu's) and giant cell arteritis and neurofibromatosis, whereas congenital aortic hypoplasia is ascribed to a developmental anomaly in the fusion and maturation of the paired embryonic dorsal aortas.[1-4] Neurofibromatosis may result in coronary, cerebrovascular, and visceral artery stenoses due to vasculopathy associated with spindle cell proliferation in the arterial wall; stenoses in the thoracic or abdominal aorta as well as the innominate and subclavian arteries have been attributed to adventitial neurofibromas.[1,3] The segmental aortic constriction of our patient was characterized by diminution of the external diameter, moderate intimal hyperplasia, and minimal periaortic inflammation. MAS is associated with a high propensity for both visceral (50%-70%) and renal (>80%) artery stenoses.[2,3] It is the consequences of renovascular hypertension, including myocardial infarction, heart failure, intracranial hemorrhage, and aortic rupture, that best portray the clinical progression of MAS if left unrepaired,[2-4] most of untreated patients succumbing before the age of 40.[2,3] The treatment of a tubular aortic narrowing (MAS), associated with renovascular hypertension and visceral artery stenosis is predominantly surgical, entailing aorto-aortic bypass of the diseased segment or, less often, aortic patching, in addition to bypass grafting of the stenosed renal and visceral arteries[1-4]; hypertension is thus improved or cured in over 70% of patients.[3] Early morbidity includes paradoxical hypertension, visceral hyperperfusion, and, rarely, spinal ischemia and paraplegia.[3]

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